General

Guideline Title

Sleep disorders in neurodegenerative disorders and stroke.

Bibliographic Source(s)

Jennum P, Santamaria Cano J, Bassetti C, Clarenbach P, Hogl B, Mathis J, Poirrier R, Sonka K, Svanborg E, Dolenc Groselj L, Kaynak D, Kruger M, Papavasiliou A, Zahariev Z. Sleep disorders in neurodegenerative disorders and stroke. In: Gilhus NE, Barnes MP, Brainin M, editor(s). European handbook of neurological management. 2nd ed. Vol. 1. Oxford (UK): Wiley-Blackwell; 2011. p. 529-43. [159 references]

Guideline Status

This is the current release of the guideline.

This guideline updates a previous version; Jennum P, Santamaria J, Members of the Task Force. Report of an EFNS task force on management of sleep disorders in neurologic disease (degenerative neurologic disorders and stroke). Eur J Neurol 2007 Nov;14(11):1189-200.

Recommendations

Major Recommendations

The levels of evidence (Class I-IV) supporting the recommendations and ratings of recommendations (A-C, Good Practice Point) are defined at the end of the "Major Recommendations" field.

Sleep Disorders Associated with Neurological Disease

Tauopathies

Sleep disorders are commonly observed in patients with tauopathies, and there should be an increased awareness of these disorders. It is recommended to perform a detailed medical history of sleep disorders in tauopathies, i.e., insomnia, excessive daytime sleepiness (EDS), motor and dreaming activity, and sleep-disordered breathing (SDB). Polysomnography (PSG) recording, preferably with audiovisual recording, is suggested for the diagnosis, especially when rapid eye movement (REM) sleep behaviour disorder (RBD) and/or SDB are suspected disorders (Level C).

Synucleinopathies

The majority of patients with synucleinopathies experience one or more sleep disorders. It is recommended to perform a detailed medical history of sleep disorders in tauopathies, i.e., insomnia, EDS, motor and dreaming activity, and SDB. PSG recording, preferably with audiovisual

recording, is suggested for the diagnosis, especially when RBD and/or SDB is suspected (Level B).

Stroke

Sleep disorders, especially SDB, occur often in stroke patients. Screening for SDB and other sleep disorders is recommended as part of the stroke evaluation programme, especially in ischaemic stroke patients (Level A).

Motor Neuron, Motor End Plate, and Muscle Diseases

SDB often occurs in patients with motor neuron, motor end plate, and muscle diseases, and should be considered in all patients. Minimum evaluation should include PSG eventually combined with additional carbon dioxide analysis, and eventually supplied with serial polygraphy or oximetry measures for the identification of sleep-related hypoventilation during the disease course (Level B).

Genetic Neurodegenerative Disorders

Sleep disorders occur in several genetic neurological diseases. The patients should be questioned, and further evaluation of these disorders should rely on a clinical judgement (Level C).

Management of Sleep Disorders in Neurological Diseases

1. Patients with neurological diseases often have significant sleep disorders that affect sleep and daytime function, with increased morbidity and even mortality. Many of these disorders are treatable. Therefore, increased awareness should be directed toward sleep disorders in patients with neurodegenerative, cerebrovascular, and neuromuscular diseases. Despite this, there are limited number of studies with a high evidence level.
2. PSG is a diagnostic minimum for the diagnoses of sleep disorders in patients with neurological diseases.
3. In patients with nocturnal motor and/behavior manifestations, a full video-PSG/video-electroencephalography (EEG)-PSG is recommended.
4. Respiratory polygraphy has a moderate sensitivity and specificity in the diagnosis of obstructive sleep apnoea syndrome (OSAS) without neurological diseases, but its value for the diagnosis of other SDBs or in neurological patients with suspected OSAS has not been evaluated compared to gold standard of PSG. Consequently, respiratory polygraphy may be used as a method for detecting OSAS, but the value of its use for SDB in patients with neurological diseases needs further validation.
5. Oximetry has a poor sensitivity/specificity for the identification of OSAS in patients without neurological diseases. Oximetry cannot differentiate between obstructive and central sleep apnoea or is insufficient to identify stridor. Oximetry alone is not recommended for the diagnosis of SDB in neurological disorders.
6. Patients with SDB, muscle weakness, and cardiac or pulmonary comorbidity may present a sleep hypoventilation syndrome that manifests early as increased carbon dioxide. Partial pressure of arterial carbon dioxide (paCO) should be measured in such cases during sleep recordings.
7. Fixed pressure continuous positive airway pressure (CPAP)/auto-adjusted CPAP is the most effective treatment of OSAS. This probably also includes patients with OSAS and neurologic diseases. However, there is a need for further evaluation of the effect of CPAP in patients with OSAS and neurologic diseases.
8. Bi-level/variable positive-airway pressure ventilation, noninvasive positive pressure ventilation (NIPPV) and volumetric ventilation is useful for SDBs such as central apnoeas, Cheyne-Stokes breathing, and alveolar hypoventilation.
9. There is a clear need for further studies focusing on the diagnostic procedures and treatment modalities in neurological patients with sleep disorders.

Definitions:

Evidence Classification Scheme for a Diagnostic Measure

Class I: A prospective study in a broad spectrum of persons with the suspected condition, using a 'gold standard' for case definition, where the test is applied in a blinded evaluation, and enabling the assessment of appropriate tests of diagnostic accuracy

Class II: A prospective study of a narrow spectrum of persons with the suspected condition, or a well-designed retrospective study of a broad spectrum of persons with an established condition (by 'gold standard') compared to a broad spectrum of controls, where test is applied in a blinded evaluation, and enabling the assessment of appropriate tests of diagnostic accuracy

Class III: Evidence provided by a retrospective study where either persons with the established condition or controls are of a narrow spectrum, and where test is applied in a blinded evaluation

Class IV: Any design where test is not applied in blinded evaluation OR evidence provided by expert opinion alone or in descriptive case series

(without controls)

Evidence Classification Scheme for a Therapeutic Intervention

Class I: An adequately powered prospective, randomized, controlled clinical trial with masked outcome assessment in a representative population or an adequately powered systematic review of prospective randomized controlled clinical trials with masked outcome assessment in representative populations. The following are required:

a. Randomization concealment
b. Primary outcome(s) is/are clearly defined
c. Exclusion/inclusion criteria are clearly defined
d. Adequate accounting for dropouts and crossovers with numbers sufficiently low to have minimal potential for bias
e. Relevant baseline characteristics are presented and substantially equivalent among treatment groups or there is appropriate statistical adjustment for differences

Class II: Prospective matched-group cohort study in a representative population with masked outcome assessment that meets a–e above or a randomized, controlled trial in a representative population that lacks one criteria a–e

Class III: All other controlled trials (including well-defined natural history controls or patients serving as own controls) in a representative population, where outcome assessment is independent of patient treatment

Class IV: Evidence from uncontrolled studies, case series, case reports, or expert opinion

Rating of Recommendations for a Diagnostic Measure

Level A rating (established as useful/predictive or not useful/predictive) requires at least one convincing class I study or at least two consistent, convincing class II studies.

Level B rating (established as probably useful/predictive or not useful/predictive) requires at least one convincing class II study or overwhelming class III evidence.

Level C rating (established as possibly useful/predictive or not useful/predictive) requires at least two convincing class III studies.

Rating of Recommendations for a Therapeutic Intervention

Level A rating (established as effective, ineffective, or harmful) requires at least one convincing class I study or at least two consistent, convincing class II studies.

Level B rating (probably effective, ineffective, or harmful) requires at least one convincing class II study or overwhelming class III evidence.

Level C rating (possibly effective, ineffective, or harmful) requires at least two convincing class III studies.

Good Practice Point Where there was a lack of evidence but consensus was clear, the task force has stated their opinion as Good Practice Points.

Clinical Algorithm(s)

None provided

Scope

Disease/Condition(s)

Sleep disorders associated with degenerative neurologic disorders and stroke, including:

- Insomnias
- Sleep-related breathing disorders
- Hypersomnias not due to a sleep-related breathing disorder
- Circadian rhythm sleep disorders

- Parasomnias
- Sleep-related movement disorders
- Isolated symptoms
- Other sleep disorders

Note: Only a selected number of the sleep disorders related to neurological diseases will be mentioned in this paper.

Guideline Category

Diagnosis

Evaluation

Management

Treatment

Clinical Specialty

Cardiology

Family Practice

Internal Medicine

Neurology

Psychology

Pulmonary Medicine

Sleep Medicine

Intended Users

Physicians

Psychologists/Non-physician Behavioral Health Clinicians

Respiratory Care Practitioners

Guideline Objective(s)

- To provide evidence-based recommendations in the management of sleep disorders associated with degenerative neurologic disorders and stroke
- To review the different sleep disorders occurring in degenerative neurologic diseases and stroke
- To review the different methods of sleep evaluation available in these patients

Target Population

Patients with sleep disorders associated with degenerative neurologic disorders and stroke

Interventions and Practices Considered

Evaluation/Diagnosis

1. Detailed medical history
2. Polysomnography (PSG) (routine, extended, or video)
3. Full electroencephalography-PSG
4. Respiratory polygraphy
5. Limited-channel polygraphy: oximetry (considered but not recommended)
6. Multiple Sleep Latency Test
7. Maintenance of Wakefulness Test

Management/Treatment

1. Fixed-pressure continuous positive airway pressure (CPAP)/auto-adjusted CPAP
2. Bi-level positive airway pressure (PAP)/variable PAP
3. Noninvasive positive pressure ventilation

Major Outcomes Considered

- Sensitivity and specificity of diagnostic tests
- Effectiveness of treatments: improvement in nocturnal respiratory abnormalities, daytime symptoms and function, and cognitive problems
- Quality of life
- Morbidity
- Mortality

Methodology

Methods Used to Collect/Select the Evidence

Searches of Electronic Databases

Description of Methods Used to Collect/Select the Evidence

The task force reviewed the pertinent literature covering three main areas:

1. Tauopathies (Alzheimer's disease [AD], progressive supranuclear palsy [PSP] and corticobasal degeneration [CBD])
2. Synucleinopathies (Parkinson's disease [PD], multiple system atrophy [MSA] and dementia with Lewy bodies [DLB])
3. Stroke, amyotrophic lateral sclerosis (ALS), myotonic dystrophy, myasthenia gravis, and spinocerebellar ataxias

Search Strategy

The literature search included PubMed and the Cochrane Database. These were searched until 2009 or over as much of this range as possible, looking for the different sleep disorders and symptoms in each of the most frequent or relevant degenerative neurologic disorders and stroke. Language of writing was restricted to European languages. Studies considered for inclusion were, when possible, randomized controlled trials of adult patients in any setting, suffering a neurodegenerative disorder (motor neuron disease, Parkinson's disease, Alzheimer's disease) or stroke. There had to be an explicit complaint of insomnia, parasomnia or hypersomnia in study participants. Observational studies were also included. Abstracts were selected by the chairmen and independently inspected by individual members of the task force; full papers were obtained where necessary.

Number of Source Documents

Not stated

Methods Used to Assess the Quality and Strength of the Evidence

Weighting According to a Rating Scheme (Scheme Given)

Rating Scheme for the Strength of the Evidence

Evidence Classification Scheme for a Diagnostic Measure

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Class IV: Any design where test is not applied in blinded evaluation OR evidence provided by expert opinion alone or in descriptive case series (without controls)

Evidence Classification Scheme for a Therapeutic Intervention

Class I: An adequately powered prospective, randomized, controlled clinical trial with masked outcome assessment in a representative population or an adequately powered systematic review of prospective randomized controlled clinical trials with masked outcome assessment in representative populations. The following are required:

a. Randomization concealment
b. Primary outcome(s) is/are clearly defined
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d. Adequate accounting for dropouts and crossovers with numbers sufficiently low to have minimal potential for bias
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Class II: Prospective matched-group cohort study in a representative population with masked outcome assessment that meets a–e above or a randomized, controlled trial in a representative population that lacks one criteria a–e

Class III: All other controlled trials (including well-defined natural history controls or patients serving as own controls) in a representative population, where outcome assessment is independent of patient treatment

Class IV: Evidence from uncontrolled studies, case series, case reports, or expert opinion

Methods Used to Analyze the Evidence

Systematic Review

Description of the Methods Used to Analyze the Evidence

A classification of the different studies according to evidence levels for therapeutic interventions and diagnostic measures was done in accordance with the European Federation of Neurological Societies (EFNS) guidance (see the "Availability of Companion Documents" field). The panel discussed what possible diagnostic tests and healthcare interventions could be recommended in each particular disease.

Method for Reaching Consensus

Where there was uncertainty further discussion was sought by the panel. Data extraction and quality assessments were undertaken independently by the panel reviewers.

Methods Used to Formulate the Recommendations

Expert Consensus

Description of Methods Used to Formulate the Recommendations

Not stated

Rating Scheme for the Strength of the Recommendations

Rating of Recommendations for a Diagnostic Measure

Level A rating (established as useful/predictive or not useful/predictive) requires at least one convincing class I study or at least two consistent, convincing class II studies.

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Good Practice Point Where there was a lack of evidence but consensus was clear, the task force has stated their opinion as Good Practice Points.

Cost Analysis

A formal cost analysis was not performed and published cost analyses were not reviewed.

Method of Guideline Validation

Peer Review

Description of Method of Guideline Validation

The guidelines were validated according to the European Federation of Neurological Societies (EFNS) criteria (see the "Availability of Companion Documents" field).

Evidence Supporting the Recommendations

Type of Evidence Supporting the Recommendations

The type of supporting evidence is identified and graded for selected recommendations (see the "Major Recommendations" field).

Benefits/Harms of Implementing the Guideline Recommendations

Potential Benefits

Appropriate diagnosis and management of sleep disorders in patients with degenerative neurologic disorders and stroke

Potential Harms

Not stated

Qualifying Statements

Qualifying Statements

This guideline provides the view of an expert task force appointed by the Scientific Committee of the European Federation of Neurological Societies (EFNS). It represents a peer-reviewed statement of minimum desirable standards for the guidance of practice based on the best available evidence. It is not intended to have legally binding implications in individual cases.

Implementation of the Guideline

Description of Implementation Strategy

The European Federation of Neurological Societies has a mailing list and all guideline papers go to national societies, national ministries of health, World Health Organisation, European Union, and a number of other destinations. Corporate support is recruited to buy large numbers of reprints of the guideline papers and permission is given to sponsoring companies to distribute the guideline papers from their commercial channels, provided there is no advertising attached.

Institute of Medicine (IOM) National Healthcare Quality Report Categories

IOM Care Need

Getting Better

Living with Illness

IOM Domain

Effectiveness

Identifying Information and Availability

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Adaptation

Not applicable: The guideline was not adapted from another source.

Date Released

2007 Nov (revised 2011)

Guideline Developer(s)

European Academy of Neurology - Medical Specialty Society

Source(s) of Funding

European Federation of Neurological Societies

Guideline Committee

European Federation of Neurological Societies Task Force on Sleep Disorders in Neurodegenerative Disorders and Stroke

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Financial Disclosures/Conflicts of Interest

None reported

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Guideline Availability

Electronic copies: Available in Portable Document Format (PDF) from the European Federation of Neurological Societies (EFNS) Web site .

Availability of Companion Documents

The following is available:

- Brainin M, Barnes M, Baron JC, Gilhus NE, Hughes R, Selmaj K, Waldemar G; Guideline Standards Subcommittee of the EFNS Scientific

Committee. Guidance for the preparation of neurological management guidelines by EFNS scientific task forces – revised recommendations 2004. Eur J Neurol. 2004 Sep;11(9):577-81. Electronic copies: Available in Portable Document Format (PDF) from the European Federation of Neurological Societies Web site .

Patient Resources

None available

NGC Status

This NGC summary was completed by ECRI Institute on April 24, 2009. This NGC summary was updated by ECRI Institute on February 20, 2012.

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